Symptom Control and End of Life Care

MND Special Interest Groups Workshop
Dr Cynthia Parr
Staff Specialist in Palliative Medicine
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"The best test of a physician's suitability for the specialized practice of neurology is not his ability to memorize improbable syndromes but whether he can continue to support a case of motor neurone disease..."

- Bryan Matthews
"Practical Neurology"

Patrick Joyce-artist

Multiple Teams

- Neurology
- Rehabilitation
- Primary Care
- Respiratory
  - ventilatory support
- Gastroenterology
  - PEG
- Palliative Care

Palliative Care

- An approach that improves the quality of life of patients and their families facing problems associated with life-threatening illness, through the prevention and relief of suffering by means of early identification and impeccable assessment and treatment of pain and other problems, physical, psychosocial and spiritual
  - WHO 2002

Palliative approach

- **Ensures**
  - early discussion about future care management decisions and advanced care planning
  - optimal symptom and wellbeing needs for the person with MND and their carer/family
- **Aims to**
  - Assist people with MND to maintain QOL
  - Support MND carers to maintain their own health

From the time of diagnosis with MND it is important to adopt a palliative approach to all aspects of the disease

Symptoms

- Dysphagia 87%
- Dyspnoea 85%
- Weight Loss 84%
- Speech Problems 74%
- Pain 73%
- Constipation 53%
- Drooling 23%
- Emotional lability 23%

Oliver 1996

Dysphagia

- Weakened muscles in chewing, moving bolus of food and swallowing

Dysphagia

- Careful feeding
  - positioning
  - consistency of food
  - small, frequent meals
- Time for eating
- Support of family
- Involvement of speech and language therapist and dietitian
- PEG feeding

PEG feeding

- **Indications**
  - acceleration of weight loss with dysphagia
  - frequent choking
  - exhaustion with eating
  - impaired QOL

PEG tube

- Best done while the respiratory function allows for a safe anaesthetic
  FVC > 50% of expected
- Once PEG inserted can use it whenever required
- Can combine oral and PEG feeds

Pharmacological measures

- With worsening dyspnoea may need to use small regular doses of morphine and lorazepam
- Used appropriately opioids and BZDs will not hasten death

Opioids in MND

- Oral morphine

mean dose	96mg/24 hours
median dose	60mg/24 hours
mean duration of use	95 days

Impaired communication

- speech therapy
- communication aids
  - bells, buzzer
  - communication boards
  - Canon communicators and lightwriters
  - picture and letter boards
  - voice amplifiers
  - computerised speech devices
  - iPads/iPhones

Pain

- Musculoskeletal
  - Joint pain
  - Stiffness
- Muscle Cramp
- Skin Pressure Pain
  - Discomfort
  - Position unchanged

Pain

- mattress support
- physiotherapy, ROM exercises, positioning
- NSAID
- quinine, carbamezepine, magnesium for cramping
- baclofen, BZD, gabapentin, cannabinoids for spasticity
- opioids for skin pressure pain

Limb weakness

- Physiotherapy crucial
- Regular exercise of muscle groups
- Orthotics
- Aids for mobilization-walking sticks to wheelchairs

Sexuality

- Most ALS patients preserve sexual function

Constipation

- coloxyl and senna
- daily microlax enema
- high fibre and fluid diet

Secretions

- Pooling of saliva
- Reduced swallowing of normal saliva
- Tenacious, thick secretions
- Secondary to sialorrhoea, dehydration and mouth breathing

Secretions

- If a patient has simultaneously thick secretions and respiratory dysfunction, the combination may lead to increasing difficulties coughing up secretions which may lead to panic and further respiratory distress

Tenacious Secretions

- Frozen dark grape juice to suck
- Normal saline nebs
- Mucomyst (N-acetyl cysteine) 3-4 mL tds-qid

Dry mouth

- Add water to meals
- Check meds
- Avoid dairy products
- Artificial saliva
- Diluted lemon juice
- Pilocarpine mouth wash
- Biotene mouth products

Secretions

- **anticholinergic medication:**
  - tricyclic antidepressants
  - hyoscine hydrobromide
  - glycopyrrolate
  - atropine
  - ipatroprium
  - scopolamine

Secretions

- clear secretions
  - manual suction, chest physiotherapy with posterior drainage, vibrating chest vest, breathing techniques, nebs, cough assist device
- botulinum A toxin injections
- radiotherapy
- surgery

Miscellaneous

- Reflux
- Dependent oedema
- Urinary urgency

Issues with interventions

- **Reduced discussion of future**
  - Disease progression
  - Effectiveness of intervention
  - Discussion of dying
  - Preparation
    - family
  - Advance care planning
    - Advanced directives
    - Place of death
    - Place of care

Emotional Lability and Dementia

- **Emotional lability** in up to 50% patients with ALS, probably more common in bulbar disease
  - not a mood disorder
  - Rx
    - amitriptylline
    - fluvoxamine
    - dextromorphan
    - quinidine
- **Fronto-temporal dementia** 5-10%
- **Cognitive impairment** 60% which interferes with planning, executive function and capacity to accept change

Psychosocial/spiritual issues

- **Depression 5-75%**
  - Prevalence not well defined, clinically major depression in 9%
  - no association with severity of illness
  - Rx psychotherapy, spiritual and social support, anti-depressants
    - amitriptyline 25 mg- 150 mg nocte
    - If depression combined with emotional lability consider cipramil/efexor

Patient concerns

- How will I be cared for?
- Will I be a burden on my family?
- Do I go to hospital or a hospice?
- Can I stay at home until I die?
- How will I actually die?

Family concerns

- Diagnosis
- Communication-physical problems, interpersonal communication
- Isolation
- Sexuality and intimacy
- Finances
- Death and dying

Spiritual aspects

- Not necessarily religious
- The meaning of life
- Why me?
- Fears of dying
- Fears of death

Fears of dying

- Choking
- Pain
- Dyspnoea

Terminal phase

- Most patients die of respiratory failure (85%)
  - Neuvert C, Oliver D et al J of Neurology 2001;248:612-616

End of life issues

- most patients died peacefully
- most common symptoms were dyspnoea, anxiety, cough
- home death feasible 50%
- main palliative measures were PEG, NIPPV, morphine, BZD
- **NONE** died choking
  - Neudert 2001

End of life care

- Anticipation
- Symptom control
- Communication is key
  - –Patient
  - –Family
  - –Carers

Planning for end of life care

- **Diagnosis**
  - – No curative treatment
  - – Palliative care from diagnosis
- **Decision points**
  - – interventions
- **Crises**
  - – deterioration
- **Terminal stages**

Planning for end of life care

- **Decision making**
  - – Resuscitation
  - – Sedation
  - – Hydration/feeding
- **End of life discussions**
  - – Euthanasia
  - – Is death discussed

Terminal Phase

- Deterioration over a few days or hours
- Often preceded by
  - not wanting to eat or dink
  - not wanting to engage
  - increased sleepiness and weakness
  - reduction in chest expansion
  - breathing irregular, rattly
  - colour changes in hands and feet
  - skin temperature becoming cold

Preparing the family

- To help prevent the “unexpected” or family shock
- Explain what signs to expect
- How and who to contact for immediate advice
- How to involve others
- Additional comfort items
- Managing the moments following death

Supporting the family

- Share care plans
- Adequate nursing cover
- Comprehensive symptom control
- Psychosocial support
- Enlist family and friends to support primary carer

End of Life Medications

- Subcutaneous route
- Morphine
- Midazolam
- Glycopyrrolate

- Withdrawal of equipment and unnecessary interventions

Self-care

- An intense time for families and health care professionals
- Important to recognise and acknowledge the impact on yourself

Bereavement

- The patient's deterioration and death will be remembered forever by the family
- Be honest, sensitive, empathic, understandable and direct with a constant balance of hope with realism
- Be aware of own need for support and feelings to prevent compassion fatigue

Conclusions

- Palliative approach from time of diagnosis and multidisciplinary care is optimal
- Complications need to be anticipated, discussed and planned for
- Family and caregivers need to be included in all aspects of care