

Motor Neurone Disease Amyotrophic Lateral Sclerosis

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ALS/MND

ALS/MND is a relentlessly progressive neurological disorder culminating in complete dependence on others for all activities of daily living.

The clinical management of ALS/MND is palliative from the time of diagnosis and is focused on symptom control and adjustment to the progressive loss of neurological function with the certainty of early death.

ALS/MND

What began as a minor symptom has turned into a disabling and fatal illness for which there is no cure.

ALS/MND

In Ireland one person dies from ALS/MND every four days.

ALS/MND

The impact of ALS/MND is a unique and individualised experience. Influenced by how the patient and their carer perceive the changes caused by the illness, their previous coping mechanisms, the presence of support structures and the strength of their relationship prior to the diagnosis.

ALS/MND is a series of losses

- **Control, independence, security**
- **Identity, role, status, self esteem**
- **Body image, sense of attractiveness, opportunities for privacy and intimacy**
- **Friends, work colleagues, extended family members**
- **Future plans and ambitions**

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For many patients with ALS/MND the shock of the diagnosis is compounded by the relentless progression of the illness. Some continue to maintain hope and a positive outlook and report to having good quality of life, living for today.

“As one finds meaning in the present, it is possible for life to be experienced as deeper, richer and more rewarding even while living with physical decline, realizing one’s finitude, the present time becomes more precious”

(Lambert 2006)

The Diagnosis of ALS/MND

Challenges a Person:

- to unlearn mastery of the body,
- to unlearn personal control and independence,
- to return to a state of complete dependence.

D Gelinas, R. Miller

Clinical features of ALS/MND that relate to prognosis

- **Cognitive Impairment**
- **Bulbar v Spinal**
- **Age of Onset**
- **Rate of Disease Progression**
- **Respiratory function**
- **Nutritional Status**

Management of ALS/MND

Symptom management

MDT: Close collaboration with PC and Resp teams

Disease Modification - Riluzole

Palliative Care: home care, day care, respite.

Management of ALS/MND

- **Improve Symptoms,**
- **Anticipate needs of patients and carers,**
- **Avoid Crisis,**
- **Adaptation of environment to promote ADL,**
- **Maintain Independence,**
- **Promote Autonomy,**
- **Ensure Dignity,**
- **Enhance Quality of life.**

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Nutritional decline can occur in the context of evolving dysphagia:

Weight loss may also be due to hypermetabolism, particularly in those with respiratory compromise.

Treatment:

Modification of food and fluid consistency, postural advice and parenteral feeding by gastrostomy.

PEG-RIG

ALS/MND

Dysarthria: Dysphagia

The ability to communicate is a fundamental human activity.

Bulbar impairment is one of the most important clinical problems because it impacts on swallowing and communication.

The impairment may range from mild dysarthria to anarthria.

Cognitive and language impairment: deficits of verbal fluency and spelling.

ALS/MND

Speech as part of communication conveys personality, such as sense of humour.

- **Isolation**
- **Frustration**
- **Increased fear and anxiety**
- **Low self esteem**
- **Loss of control**
- **Increased sadness**

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Bulbar dysfunction and related communication, secretion and respiratory complications are major issues and impact on quality of life.

Communication:

The goal should be to optimize the communication both for patient and the carer. AAC devices can be used in those with intact cognition.

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Sialorrhoea and Bronchial Secretions:

Problems with oro-pharyngeal secretions often co-occur with speech difficulties and usually with impairment of swallowing.

Sialorrhoea is very distressing to patients, and increases the risk of oral infections and can be very difficult to manage.

Treatments:

Amitriptyline, Atropine, transdermal hyoscine, glycopyrrolate, salivary gland irradiation, botulinum toxin.

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Sialorrhoea and Bronchial Secretions:

Bronchial secretions can be treated with mucolytics, anticholinergics and in some instances, cough assist (insufflator-exsufflator).

Bach has long been a strong proponent of cough assisting techniques and mechanical devices to aid in effective airway management.

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CoughAssist Mechanical Insufflator-Exsufflator:

The coughassist is a non-invasive therapy that safely and consistently removes secretions in patients with an ineffective ability to cough.

Benefits of coughassist:

- **Removes secretion from the lungs**
- **Reduces the occurrence of respiratory infection**
- **Safe, non-invasive alternative to suctioning**
- **Easy for patients and caregivers to operate**

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Respiratory Insufficiency:

The majority of patients die from respiratory failure, and the presence of respiratory muscle weakness is an independent predictor of quality of life.

Symptoms:

- **Breathlessness**
- **Early morning headaches**
- **Daytime sleepiness, nocturnal agitation**
- **Nightmares**
- **Confusion**

Recognition, early intervention

Management

- **End of life decisions**
- **Non invasive ventilation**
- **Invasive mechanical ventilation**

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Non-invasive ventilation:

Patients attending a specialist ALS/MND clinic are more likely to undergo routine respiratory monitoring and to be successfully established on NIV.

Initiation of NIV is associated with a reduction in the initiation of NIV in an emergency situation and the use of invasive ventilation.

Limitations of NIV:

Cognitive impairment

Bulbar function

Non-Invasive Ventilation (NIV)

Criteria for initiation of NIV in ALS

- Symptoms of hypoxemia
- FVC <50%, SNIP < 40cm H₂O
- Pulse oximetry
- Elevated early morning ABG
- NIV extends survival (> 5hrs)
- Oxygen (decreases resp drive)
- PAV
- Locked in Syndrome

- End of Life issues

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Non-invasive ventilation:

Cost : NIV = Cough Assist = 650 Euro per month Respi Care

Patients can survive for many years with 24 hour NIV particularly when techniques are used to achieve adequate management of secretions

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Non-invasive ventilation:

NIV is now a standard of care in ALS/MND,

However, there is marked variation in the use of NIV within and between countries.

NIV:

- **Reduces dyspnoea**
- **Improves quality of life**
- **Prolongs survival**

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Invasive ventilation:

Tracheostomy with invasive ventilation: (TV)

The use of TV remains highly controversial, even in those countries where it is most widely utilized. It is not routinely available outside the US and Japan due to the financial burden it represents to individuals and society as a whole.

Europe = 0% - 10.6%

US = 2% - 6%

Japan = 25% 46%

Cost: 300,000 – 400,000 euro per annum

ALS/MND

Invasive ventilation:

Survival after TV varies widely, from months to several years. The illness continues to progress to a point where some patients become “locked in”, a state in which they cannot communicate but breathing is maintained indefinitely.

TV poses many challenges:

It is viable and desirable option for some patients and the majority of these patients report contentment and happiness with their choice.

Family and caregivers felt TV was a major burden that their own health had suffered, they reported a loss of privacy in the home as well as increased tension, depression and anxiety.

Dilemmas

“ALS/MND has been identified as an illness that is particularly prone to engendering a sense of helplessness and failure which leads to feelings of frustration and de-skilling that can result in health-care professionals avoiding patients”.

(Skyes 2006)

“The care of people with ALS/MND can raise difficult ethical dilemmas, shifting from a paradigm of cure to one of symptom management and palliation can be difficult for health-care professionals in acute hospitals”

(Oliver et al. 2004)

ALS/MND

Ethical Dilemmas:

PEG - RIG

Patient Autonomy:

Patient Autonomy

“Autonomy is not just a status, but a skill, one that must be developed. Health-care interactions rely upon assent, rather than upon genuinely autonomous consent. Throughout most of their lives, patients are socialised to be heteronomous, rather than autonomous.

At the worst possible time – when life and death consequences are attached to choices, the paradigm shifts and real consent is sought, even demanded, making an often traumatic situation even harder”
(C. Myers 2004)

Patient Autonomy

Patients need to be aware that their autonomy is not absolute, that they cannot insist on receiving specific, unrealistic or illegal treatment.

Patients need to understand the implications of their decisions. That their decisions may influence the treatments they receive, the services that are available to them and may determine their place of death.

End of Life Decisions

As treatments are limited, inevitable decision regarding accepting or forgoing life-sustaining therapies should be made. The failure to address advance care planning leads to unplanned interventions, particularly mechanical ventilation.

An acknowledgement by the patient, at some level, that death, while not imminent, will be the likely outcome of the disease.

Advance Care Directive:

Any statement made by a competent individual with the intention of exerting control over medical care at such time that the individual is unable to make or express decisions regarding such care.

End of Life Decisions

Advance Care Directives are increasingly recognised as an important tool for safeguarding patients' autonomy.

However, my study identified that in an Irish context the use of ACD's cannot promise or guarantee patients with ALS/MND a say in their future care and may not ameliorate quality of life in the terminal phase of the illness. It has identified that patients with ALS/MND who wish to invoke an ACD are completely dependent on the attending physician.

Health-care professionals have a right to their autonomy and the right to care for individuals without abandoning their own integrity.

Patient Autonomy

Maintaining control is vital for ALS/MND patients

- **Locus of Care**
- **Family/Carer support (24/7)**
- **Financial Burden**
- **Fully informed**
- **Locked in Syndrome**

- **Is the patient ever truly Autonomous!!**

Difficulties in Caring for patients with ALS/MND

- **Unpredictability of the course of the illness!**
- **Heavy burden of care!**
- **ALS/MND patients are very demanding!**
- **Ethical issues regarding end of life decisions!**

Terminal Phase

Medication in last days of Life: (Oliver et al 2009).

60 patients included: 63% male/37% female

Mean time from first symptom to death was 32 months

The majority of patients received medication in the last 72 hours of life:

- **Morphine- 23 pts in last 24hrs, commonly (38%) by subcut with a mean dose of 80mgs (oral equivalent) over 24hrs**
- **Midazolam- 35 pts, commonly sub/cut with a mean dose of 31mg/24 hours**
- **Anticholinergic medication – glycopyrronium bromide or hyoscine hydrobromide – 35 pts**

All patients reported as dying peacefully without distress.

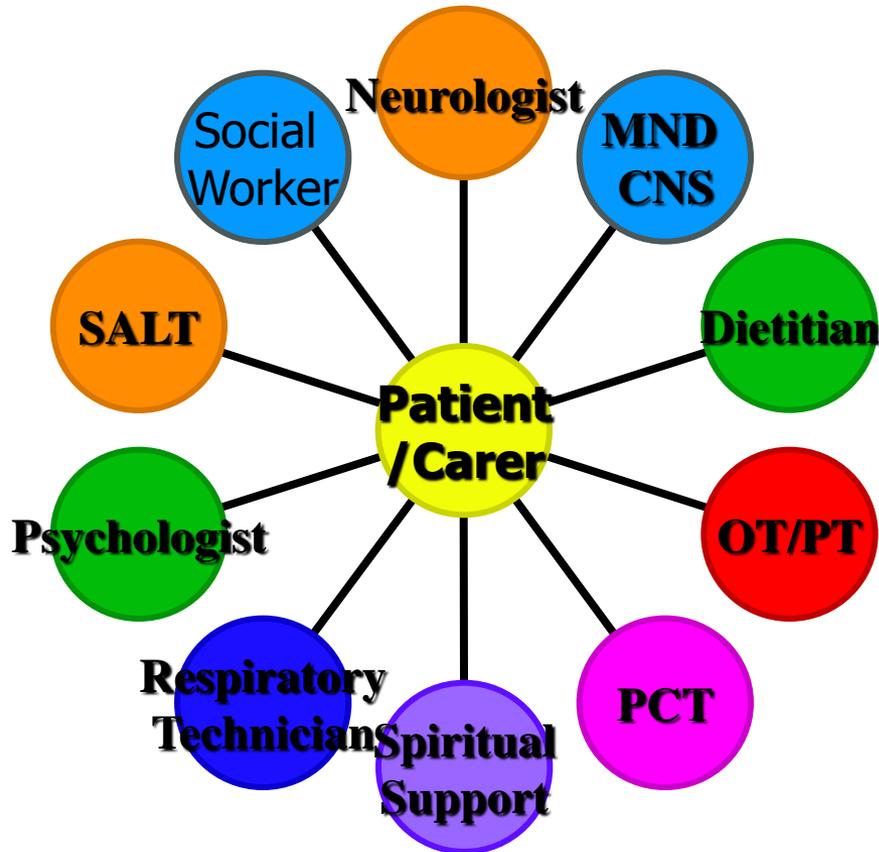
Results: HCP can feel secure in the administration of medication at the E-o-L, doses are not large.

Patients and families can be reassured that with good symptom management, and the best use of medication dying from ALS/MND can be peaceful!

Management of ALS/MND

- **Team approach**
- **Early referral**
- **Prompt intervention**
- **Flexibility**
- **Frequent assessment**
- **Multidisciplinary team**

Multidisciplinary Management



Conclusions

ALS/MND is a complex disorder

- **Disease modifying therapies limited**
- **Early intervention can alleviate symptoms**
- **Multidisciplinary care can enhance quality of life and improve survival**

Conclusion

As HCP we face many complex and ethical dilemmas regarding the management and treatment interventions in caring for these vulnerable patients and their families. We need to accept the limitations of the care that can be given and approach patients and their families in a positive, but realistic way.

We have a responsibility to be honest regarding the appropriateness of these treatments, the benefits and the risks, and the realistic choices available need to be discussed.

We need to ensure that the patient and their family are fully informed about the likely disease outcome, and the consequences of their choices.

Conclusion

The treatments that can currently be offered for people with Motor Neurone Disease will at best delay the progression of the disease process, are never curative so that the care of these patients is palliative from the time of diagnosis”

David Oliver, Wisdom Hospice

In caring for MND/ALS patients we should aspire to achieve similar outcomes as reported by Ganzini (2002), that most ALS/MND patients anticipate and plan for their deaths and have their wishes respected!