Occupational Therapy

Motor neurone disease / Amyotrophic lateral sclerosis
MND / ALS

- degenerative disease of upper and motor neurons of CNS
- progressive paralysis of all voluntary muscles
- no change in sensation
- Cognitive deficits in fronto-temporal dysfunction in some cases

Nervous system
Different terms / names
- motor neurone disease (UK & commonwealth)
- amyotrophic lateral sclerosis (continent)
- Lou Gerigs Disease (USA)

Epidemiology
- incidence 1.5-2.0 per 100,000 per year
- prevalence 6-8 per 100,000
- gender 1.5:1 M:F
- average age onset 58yrs

Different types
- Amyotrophic lateral sclerosis
- primary lateral sclerosis
- progressive muscular atrophy
- progressive bulbar palsy
- pseudobulbar palsy
- 'Flail arm' variant
- ALS / FTD (dementia)

Clinical course
- constant progression
- different speeds depending on type of onset
- no remissions
- median life expectancy: 3-4 years spinal onset, 1-2 years bulbar onset, <10% >6/7 years
### ALS: a progressive illness

- Loss of mobility
- Loss of manual dexterity
- Loss of writing ability
- Loss of driving ability
- Loss of self care ability
- Loss of swallow
- Loss of speech
- Loss of breathing ability
- Loss of social role

### ALS: a progressive illness

- Loss of family role
- Loss of faith
- Loss of hope
- Loss of meaning in life
- Loss of independence
- Loss of emotional control

### Symptoms of ALS

**Direct**
- Weakness (UMN & LMN) and muscle atrophy (LMN)
- Fasciculations (LMN)
- Spasticity (UMN)
- Dysarthria (UMN & LMN)
- Dysphagia (UMN & LMN)
- Dyspnoea (LMN)
- Emotional lability – pathological laughing / crying (UMN)
- Cognition (fronto-temporal cortex)

**Indirect**
- Psychological problems
- Pain
- Sleep disturbance
- Constipation
- Hypoventilation
- Musculoskeletal change
- Drooling
- Panic
- Anxiety
Limb and bulbar effects

Thinking (cognitive) and/or Behavioural problems can occur in MND

- Some patients with MND appear to be completely unaffected by these problems
- Some patients only develop limited cognitive and/or behavioural changes
- Some patients develop a more serious form of these problems – a type of dementia called Frontotemporal Lobar Degeneration (FTLD)

Directly affects

- Understanding OT intervention
- Compliance
- Remembering advice, contacts, links with other services
- Safety awareness
- Self judgement about mobility and function

Determines how we and to whom we provide information and advice – significant other / carer

FTLD

Degeneration of the frontal and temporal lobes of the brain.

Frontal variant
Semantic dementia
Non fluent progressive aphasia
Prevalence and patterns of cognitive impairment in sporadic ALS

G.M. Ringholz, MD, PhD; S.H. Appel, MD; M. Bradshaw, PhD; N.A. Cooke, PhD; D.M. Mosnik, PhD; and P.E. Schulz, MD

Neurology 2005;65:586-590

Conclusions: These data confirm the presence of cognitive impairment in 50% of patients with ALS and particularly implicate executive dysfunction and mild memory decline in the disease process.

More severe impairment occurs in a subset of patients with ALS and has features consistent with FTD.

Occupational performance

- Described as an individual’s ability to perform activities of daily living, influenced by the environment in order to maintain roles

- Activities of daily living are the range of every day tasks to fulfill occupational therapy roles in self care, work & leisure

Consider

- Person’s individual needs
- Type of ALS
- Environment
- Person’s goals, choice, autonomy
- Age
- Carers / significant other
- Progressive nature of MND / ALS
- Palliative approach

Performance components

- Motor i.e. strength, ROM, tone, loss of endurance, posture, dexterity
- Cognition i.e. executive skills; awareness, problem solving, understanding, insight, personality, behavioural change, emotional lability
- Psychological i.e. individual coping patterns
- Social i.e. supports, social participation
All components affect:
- Independence in;
- Mobility
- Personal care
- Transfers
- Driving
- Home activities
- Social activities

OT evidence
- Little published OT evidence
- Qualitative study on occupational performance (Brott et al. 2007)… occupational disruption early identification of illness….occupational performance compromised by disability….occupations gives meaning to life, form identity and occupy time….role loss….effect of assistive devices on occupational performance….change in others’ perception of person when their occupational performance was affected.
- Research primarily on MDT effectiveness (Van den Berg et al. 2005, Ng et al. 2009)

Modalities in OT intervention to facilitate occupational therapy performance
- Aids & equipment
- Adaptive techniques for ADLs
- Seating & positioning
- Education i.e. energy conservation
- Access & home modifications
- Assistive technology
- Wheelchair mobility
- Driving

Hard casting / Splinting N/A in ALS ??????
Neck Braces: Cervical orthoses in neurological conditions

Headmaster Collar (available from OrthoCare, Murrays Medical)

Oxford collar (www.saltstechstep.co.uk)

Cervical collars for immobilisation not applicable

Aspen Collar
Philadelphia Collar

What aids are useful and when?
Think ALS type!

- Time frame i.e. rate of progression
  - Spinal onset Vs bulbar onset
  - ALS Vs PLS
  - ‘Flail arm’ variant

All of the above have different rates and pattern of disability

Think person!

- Does client wish to use aid?
- Do they understand purpose?
- Does it really make a difference?
- Judge compliance (cognition, behaviour)
- Client’s environment, supports, primary carer & their role

Think system!

- Entitlements
- Local Vs national
- IMNDA equipment bank
- Do we plan in advance of need to accommodate for HSE ‘red tape’ and delays in providing services in ALS? i.e. not all those who have ALS are diagnosed with ALS!
- How well (or not) do public and private health care mix in ALS?

OT work settings

- Acute hospital (secondary, tertiary care)
- Primary care
- Voluntary sector
- Advocacy
**Intervention should be timely**

- Predict disability within general time frame
  - i.e. Spinal onset Vs bulbar onset
  - i.e. Primary lateral sclerosis Vs Amyotrophic lateral sclerosis
- Pre-empt service need to compensate for HSE inadequacies
- Available community services to monitor progression

**Absence of OT structured care**

- No overall strategic network for OT services
- Inequity across services
- Lost within an overall demand on services
- Limited OT personnel dedicated to neurology
- Absence of a structure to monitor and provide continued care
- Limited follow through by HSE on Primary Care Teams
- Inadequate funding for aids/appliances
- Access tp primary care restricted without medical care

**public versus private health care**

Does it work in ALS?

**Better outcomes in care from tertiary ALS centres**

- Traynor et al (2003): prospective population based study; better outcome in prognosis for bulbar onset
- Van den Berg et al (2005): higher QoL in psychological & social domains and better access to aids & appliances
- Van der Steen et al (2009): negligible difference between costs of MDT versus general care
- Chio et al (2008): prospective population based study; MDT clinic - independent positive factor in survival (including spinal onset)
Entitlements
- Private healthcare no automatic access to OT primary care services
- Private hospitals without OT skill mix
- Access to primary care difficult as mean tested i.e. medical card; Implications for community OT / primary care services…..
- Long term illness card N/A in MND

Without resources?
- Better knowledge of ALS / MND disability profiles
- Clinical reasoning. Progressive disability is not ‘ticky box’
- Update on the evidence
- Client perceived wellbeing is the main concern

Non HSE Support services
- IMNDA
- IWA (driving)
- Primary medical certificate & exemptions under scheme
- EU disabled parking disc
- Clients Technical Division CRC
- Local Housing authorities
- Carers Association
Case study

- 29 year old male
- Spinal onset
- Diagnosed 2009
- Initial symptoms in 2008 (falling due to right foot drop), wasting of intrinsic muscles of both hands. Increased tone in all limbs. Recent mild slurring (spastic dysarthria) with pseudobulbar affect
- Lives with spouse & 2 children
- Semi-D suburbia south Dublin
- Secondary school teacher. Recently on indefinite sick leave due to emotional lability
- Drives
- Good immediate family support
- VHI (initial diagnosis in Beacon) with referral to public ALS clinic for ongoing care

Case study

- 42 yr old female
- Diagnosed 2005, remains primarily UMN 2009
- Continues to work as solicitor part time
- Increased tone all limbs with UMN weakness, no muscle wasting
- Refused medical card on 3 applications, application on ‘compassionate ground’ also refused
- Mother of 3 under 10 years
- Lives in 3 storey town house (unwilling to modify home or accept aids beyond walking stick)
- Drives
- Spouse works fulltime
- Has private home help

Case study

- 71 year old female just received diagnosis
- Anarthric at time of diagnosis, breathing problems mid 2009
- Remains ambulant and dexterity adequate for ADLS despite mild intrinsic wasting
- Priority hospital admission for NIPPI and RIG (pending respiratory tests)
- Lives alone in North Dublin catchment area, widow
- Daughter lives nearby
- Continues to drive
- Social activities due to anarthria
- Has medical card
Thank you for participating

References

- Foley G (2007). What are the care needs for people with motor neurone disease and how can occupational therapists respond to meet these needs. British Journal of Occupational Therapy, 70(1), 32-34
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