Institut et hôpital neurologiques de Montréal
Montreal Neurological Institute and Hospital

Symptomatic management of ALS
Gestion des symptômes de la SLA

ALS Education Day

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McGill | Centre universitaire de santé McGill | McGill University Health Centre

Outline

- Multidisciplinary clinics
- ALS Symptom Management
  - *Cramps - Fasciculations*
  - Spasticity
  - Secretions
  - Pseudobulbar affect
  - Mobility and speech assistive devices
  - *Other:*
    - *Pain – Constipation – Depression – Fatigue*
- Nutritional care and dysphagia
- Respiratory care
- Palliative care
- Conclusion

MNI ALS Team

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Multidisciplinary Clinics

- Improved QOL
- Improved survival in most studies
  - Independent of the use of Riluzole – NIV – Gastrostomy
- Reduction in hospitalisations

- Longer travel time (less convenient)
- Very long tiring days
  - Depressing

Symptom management

Supportive and symptomatic management of amyotrophic lateral sclerosis Esther V. Hobson and Christopher J. McDermott. Nature Neurology Reviews, volume 12, September 2016, 526-538.

A survey of clinicians' practice in the symptomatic treatment of ALS. Dallas A Forshew and Mark B Bromberg. ALS and other motor neuron disorders 2003 4, 258–263

Problem	Pharmacological treatments	Grade of recommendation	Nonpharmacological strategies	Grade of recommendation
Cramps	Quinine[20,22]	Grade B	Physical therapy/ exercise	Grade D
	Levetiracetam[23]	Grade C		
	Mexiletine[24]	Grade B		
Spasticity	Baclofen, tizanidine or dantrolene	Grade D	Prescribed exercise[27]	Grade B
	Benzodiazepines	Grade D	Hydrotherapy	Grade D
	Intrathecal baclofen[28–30]	Grade C	Cryotherapy	Grade D
Pain	WHO Analgesic Ladder	Grade D	NA	NA
	Opioids	Grade C		
Emotional lability	SSRIs[48]	Grade C	NA	NA
	Dextromethorphan–quinidine combination treatment[46,47]	Grade A		
	Amitriptyline[49]	Grade C		
Depression	SSRIs	Grade D	Psychological therapy/ counselling	Grade D
Anxiety	Benzodiazepines	Grade D	Psychological therapy/ counselling	Grade D
Fatigue	Modafinil[53]	Grade B	NA	NA
Respiratory failure	NA	NA	Noninvasive ventilation[79]	Grade A
			Cough augmentation	Grade D
Excessive respiratory secretions (thick)	Carbocysteine	Grade D	Cough augmentation	Grade D
	Nebulised saline	Grade D	Humidification of NIV	Grade D
			Suction	Grade D
			Reduction in dairy product intake	None
			Pineapple juice	None
Excessive oral secretions (thin)	Hyoscine patches	Grade D	Radiotherapy[87,88]	Grade C
	Amitriptyline	Grade D	Suction	Grade D
	Atropine drops	Grade D		
	Glycopyrrolate	Grade D		
	Botulinum toxin[82–84]	Grade B		
Dry mouth	Artificial saliva sprays or salivation-stimulating tablets	None	Humidification of NIV	Grade D

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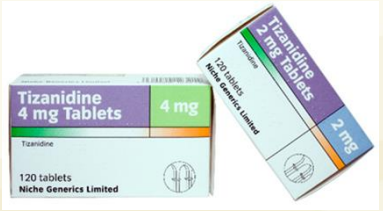
Spasticity

- Physiotherapy exercises – stretching
- Baclofen
- Tizanidine



- Marijuana

- Dantrolene
- Intrathecal Baclofen

Secretions - Saliva

- Anticholinergic medications
  - Local: Atropine drops
  - Systemic patch: Scopolamine patch
  - Oral: Amitriptyline – Glycopyrrolate
- Botulinium Toxin Injection
  - Only proven treatment by RCT
  - Small risk of increased dysphagia
- Marijuana
- Radiotherapy
- Suction device

Secretions - Thick

- Mucolytic agents
  - Carbocysteine – N/A in Canada or US
  - AcetylCysteine
- Respiratory techniques
  - Breath-stacking exercies
  - Cough Assist device (mechanical insufflation/exsufflation devices)
  - NIV (BiPap)
  - Increasing bipap humidity - Nebulised Saline
- Suction

Pseudobulbar affect
Emotional lability

- Antidepressant medications
  - TCA's: Amitriptyline
  - SSRI's: Citalopram, Escitalopram, etc…
- Dextrometharphan – Quinidine (Nudexta)

Assistive devices

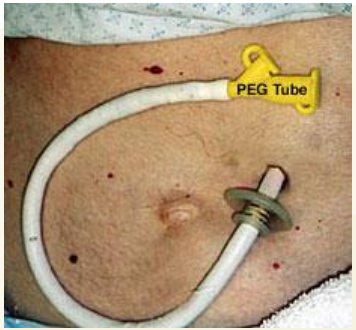
- Mobility
  - AFO and Wrist splints
  - Cane – Walker – Déambulateur
  - Wheelchairs
  - Neck collars – Thoracic/Abdminal Corset
  - Hospital Bed – Cushions (Ulcer prevention)
  - Home adaptation (Toilet access – single floor – …)
- Speech
  - Writing boards
  - Text-to-speech Apps (via tablet) – Voice banking
  - Eye-gaze software
  - Brain-Computer interfaces

Nutrition and dysphagia

- Weight loss
  - Decreased intake: dysphagia – poor appetite – limb weakness
  - Increased loss: Increased Energy needed – Muscle wasting
  - Poor prognostic factor
  - High-Calorie diet may improve survival
- Regular assessments and counseling
  - Swallowing – Techniques to prevent aspiration/choking
  - Nutritional – Change of texture, hypercaloric nutrition

Hypercaloric enteral nutrition in patients with amyotrophic lateral sclerosis: a randomised, double-blind, placebo-controlled phase 2 trial. Lancet. 2014 Jun 14;383(9934):2065-2072

Gastrostomy



- 2 Main techniques in Canada
  - Radiological guidance versus Endoscopic
- Advantages
  - Weight-stabilisation if < 10% weight loss
    - Earlier insertion
  - Possible improved survival
    - Bulbar-onset patients with dysphagia
  - Possible improved quality-of-life
    - Decreased anxiety/fatigue with meals
    - Increased care-giver burden
- Futile in frail/end-of-life patients

ProGas. Lancet Neurology 14, 702. (2015)

Respiratory care

- Regular monitoring of respiratory function recommended
  - History – Measurements
- Regular immunizations
- Non-invasive Ventilation (BiPap)
  - 1st treatment to improve
    - Survival: 7-13 months – 19 months if bulbar-onset

      AND
    - QOL – even if late in disease
- Can be challenging
  - Patient perseverence
  - Professional support
  - Parallel treatment of dry mouth – claustrophobia
- Role of opioids to alleviate dyspnea

Bourke et al. Effects of NIV on on survival and QOL in patients with ALS: a RCT. Lancet Neurol 5, 140-147 (2006).
Identifying who will benefit from NIV in ALS in a clinical cohort. Berlowitz DJ, et al. J Neurol Neurosurg Psychiatry 2016;87:280–286.

Palliative care

- Early and continuous involvement recommended
- Role in symptom control
- Alternative to Gastrostomy
- When/how to stop NIV
- End-of-life care at home or institution

- Aide Médicale à mourir

Conclusion

- Symptom management in ALS is what we do at every clinic visit

- Importance of multidisciplinary approach with
  - Other medical specialties
  - Other health professionals

- Keeping patient concerns and needs at the center of patient care

Merci!

Questions?

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