



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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Conflict of interest

- Receiving honorarium from Mitsubishi Pharma as organizer and lecturer today

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
- *Cognitive dysfunction*
- *Respiratory care*
- *Palliative care*
- Conclusion

MNI ALS Team



MNI ALS Team 2019



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		
	Intrathecal baclofen ²⁵⁻³⁰	Grade C		
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 ⁵¹	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 ^{77,88}	Grade C
	Amitriptyline	Grade D	Suction	Grade D
	Atropine drops	Grade D		
	Glycopyrrolate	Grade D		
	Botulinum toxin ⁹²⁻⁹⁴	Grade B		
Dry mouth	Artificial saliva sprays or salivation-stimulating tablets	None	Humidification of NIV	Grade D

Cramps / fasciculations

- Quinine
 - Black box warning - Katzberg et al AAN guidelines
 - Dose-dependent side-effects...
- Mexiletene
 - RCT evidence of efficacy – not best tolerance
- Others
 - Baclofen – Gabapentin/Pregabalin – Mg – Tonic water – Cyclobenzaprine

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 exercises
 - Cough Assist device (mechanical insufflation/exsufflation devices)
 - NIV (BiPap)
 - Increasing bipap humidity - Nebulised Saline
- Expectorant = Guaifenesin
- Suction



Pseudobulbar affect

Emotional lability

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

Assistive devices

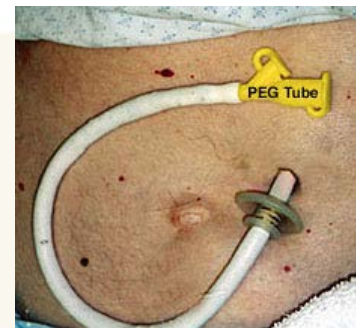
- Mobility
 - AFO and Wrist splints
 - Cane – Walker – Déambulateur
 - Wheelchairs
 - Neck collars – Thoracic/Abdominal Corset
 - Hospital Bed – Cushions (Ulcer prevention)
 - Home adaptation (Toilet access – single floor – ...)
- Speech
 - Writing boards
 - Text-to-speech Apps (via tablet) – Voice banking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

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



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 perseverance
 - Professional support
 - Parallel treatment of dry mouth – claustrophobia
- Role of opioids to alleviate dyspnea

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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