Management of Patients with ALS

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Outline

▪ What is ALS?
▪ Implications of Cognitive Involvement
▪ Dysarthria Profile
▪ Dysphagia Presentation
▪ Conclusion
Amyotrophic Lateral Sclerosis (ALS)

- A progressive Motor Neuron Disease (MND) involving both:
  - Upper Motor Neurons (UMN) – hyperreflexia, rigidity, weakness
  - Lower Motor Neuron (LMN) – hyporeflexia, muscle cramps, fasciculations, atrophy, weakness

- Bulbar vs. Spinal
Triggers

- Genetic predisposition
- Environmental factors:
  - Trauma: surgery, physical injury, falls
  - Irritants: toxins, chemicals
  - Infections
Statistics

- 3-4:1 ♀: ♂ ratio in 20s-30s vs. 1:1 post-menopause
- Incidence: 1.5-2.5/100,000/year in US
- Prevalence: 5-7/100,000 in US
- 5-10% familial vs. 90% sporadic
- 50% of the population survive 3-5 years after established diagnosis
- Average age of onset is 53-57 years
Diagnosis

- Diagnosis by excluding other conditions with related symptoms
  - Clinical examination
  - Electrodiagnostic testing
  - Radiographic exams
  - Lab/blood work up
Disease Management

- Proactive multidisciplinary symptomatic management
  - Neuroprotection
  - Antioxidants
  - Respiratory management
  - Spasticity
  - Pseudobulbar affect
  - Nutrition
  - Sialorrhea
Cognition

- 50% show evidence of cognitive impairment
- Most common pattern is mild frontal lobe executive dysfunction
- 15% of the affected patients develop frontotemporal dementia (FTD)
Cognitive Impairment

- Initiation
- Insight
- Personality
- Attention & multitasking
- Problem solving & decision making
- Organization & planning
- Perseveration & impulsivity
- Following recommendations/strategies
Cognitive Management

- Simplify
- Institute a pattern with structured schedule
- Modify the environment
- Positive reinforcement
- Encourage caregivers to change their response to the negative behavior
- Adjust roles as primary decision-maker
Dysarthria

- Speech changes typically precede dysphagia
- Type: mixed spastic (UMN) – flaccid (LMN)
- Characteristics: hypernasality, hoarseness, strain, slow rate, imprecise articulation, monopitch, reduced volume
- Bulbar onset patients had greater dysarthria at time of ALS diagnosis (Tomik & Guiloff, 2010)
- Dysarthria effects 70% of limb onset at later stages
Dysarthria Management

- Speech intelligibility strategies
- Communication strategies
- Alternative Modes of Communication:
  - No technology
  - Low technology
  - High technology
  - Apps
Dysphagia – Oral Presentation

- Initial involvement: hypoglossal > trigeminal & facial motor neurons

- Oral Dysphagia:
  - Reduced labial seal
  - Delayed bolus manipulation and transfer
  - Velopharyngeal insufficiency
  - Piece meal deglutition
Dysphagia – Pharyngeal Presentation

- Laryngeal sensory deficits found in 55% of ALS patients (Milan et al., 2006)

- Pharyngeal Dysphagia:
  - Delayed initiation of stage transition
  - Weakened pharyngeal constriction
  - Reduced hyolaryngeal excursion
  - Decreased epiglottic inversion
  - Airway invasion
Dysphagia Management

- Therapy focuses on maintaining function for as long as possible

- Management of symptoms:
  - Diet modification
  - Compensatory strategies
  - Environmental modification
  - Patient and family education
Conclusion

“There is a great deal that can be done to treat the symptoms of ALS, to improve the quality of life of the patient, and to help caregivers and the family cope with the disease”

Amyotrophic Lateral Sclerosis
A Guide for Patients and Families: Third Edition
Hiroshi Mitsumoto, MD
“Team Hope”

Thank you

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References


References


References


