ALS

ECARE SENIOR CARE 12-10-19

Objectives

- 1. Identify clinical signs and symptoms of ALS
- 2. Identify the progression of ALS
- 3. Identify symptom management of ALS

What is ALS?

- ALS stands for Amylotrophic lateral sclerosis. Also known as Lou Gehrig's disease (named after the famous baseball player who died of the disease.
- Damages the nerves that control muscles which causes the muscles to weaken.
- Leads to paralysis over time and eventually death.

Spectrum of Motor Neuron Disease

- Progressive muscular atrophy
- Primary Lateral Sclerosis
- Progressive bulbar palsy
- Flail arm syndrome
- Flail leg syndrome
- ALS-plus syndrome

Clinical Symptoms and Signs of ALS

- Initial presentation:
  - Can occur in any part of the body.
  - Usually asymmetric limb weakness is the most common presentation.

Clinical Symptoms

- Upper motor neuron symptoms
  - Slowness in movement
  - Incoordination
  - Stiffness
  - Poor dexterity
  - Spastic gait with poor balance
  - Spontaneous leg flexor spasms and ankle clonus

Clinical Symptoms

- Bulbar upper motor neuron disease
  - Dysarthria: Trouble talking
  - Dysphagia: Trouble swallowing
  - Pseudobulbar affect
  - Laryngospasm
  - Increased masseter tone
  - Stiffness
  - Imbalance

Clinical Symptoms

- Lower motor neuron symptoms
  - Atrophy
  - Fasciculations: muscle twitches
  - Muscle cramps
  - Hand weakness
    - Difficulty using buttons or zippers
    - Handling small things
    - Writing

Clinical Symptoms

- Lower motor neuron symptoms
  - Proximal arm weakness: Results in difficulty elevating the arm to the level of the mouth or above the head
    - Difficulty bathing, dressing, grooming, and eating
    - Foot and ankle weakness: Results in tripping, a slapping gait and falling

Clinical Symptoms

- Proximal leg weakness
  - Difficulty arising from chairs, climbing stairs, and getting off of the floor.
  - Impaired balance

Clinical Symptoms

- Dysarthria: may result from weakness of the tongue, lips, or palate.
- Speech is slurred and may have a nasal quality
- Hoarseness: may be caused by vocal cord weakness

- Dysphagia results from tongue weakness and disruption of the oral or phargyngeal phase of swallowing.
- Tongue weakness may result in pocketing food between cheeks and gums
- Coughing, choking on food and liquids or oral secretions
- Aspiration

Clinical Symptoms

- Lower motor neuron weakness of the upper face:
  - Incomplete eye closure
  - Poor lip seal that contributes to drooling or sialorrhea

Clinical Symptoms

- Lower motor neuron weakness affecting the trunk and spine.
  - Difficulty holding up head and difficulty maintain erect posture.

Lower motor weakness of diaphragm

- Progressive dyspnea at rest
- Reduced vocal volume
- Orthopnea and sleep disordered breathing

Clinical Symptoms

- Extraocular motor neurons: Spared until late in the disease process
  - Progressive difficulty with ocular motility
  - Locked-in state.
  - In ability to move any voluntary muscle.
  - Unable to communicate.

Clinical Symptoms

- Cognitive Symptoms:
  - Apathy
  - Loss of sympathy/empathy
  - Changes in eating behaviors
  - Disinhibition
  - Perseveration

Clinical Symptoms

- Autonomic symptoms
  - Constipation
  - Delayed colonic motility
  - Dyphagia for thin liquids related to pharyngeal muscle weakness leading to dehydration
  - Early Satiety
  - Bloating consistent with delayed gastric emptying
  - Urinary urgency without incontinence is common
  - Incontinence is uncommon

Clinical Symptoms

- Parkinsonism and supranuclear gaze palsy
  - Facial masking
  - Tremor
  - Bradykinesia
  - Postural instability

Clinical Symptoms

- Sensory symptoms
  - Tingling paresthesia
  - Pain
    - Reduced mobility: leads to skin breakdown and musculoskeletal pain
    - Muscle cramps
    - Muscle spasticity

Clinical Pattern of Progression

- Relentlessly progressive disorder with linear progression with a relatively constant slope.

- Life Threatening features:
  - Respiratory weakness and failure (most common cause of death)
  - Dysphagia
    - Risk of aspiration resulting in pneumonia
    - May lead to malnutrition and dehydration

Prognosis

- Most ALS patients die within 3-5 years of diagnosis
- Approximately 30% of ALS patients are alive five years after diagnosis
- 10-20% survive for greater than 10 years.

Multidisciplinary Care

- Neurologist
- Physical Therapist
- Occupational therapist
- Speech therapist
- Respiratory therapist
- Dietitians
- Social workers

Palliative Care

- Establishing goals of care
- Providing consistent and sustained communication between the patient and all caregivers
- Psychosocial support
- Spiritual support
- Practical support
- Coordination of care across all sites of care.

Respiratory Management

- Ventilatory support
- Immunizations
  - Seasonal influenza vaccine
  - Pneumococcal vaccine

Dysphagia and Nutrition

- Dysphagia
  - Risk of insufficient caloric and fluid intake
  - Worsening of weakness and fatigue
  - Risk of aspiration and choking

- Management
  - Modification of food and fluid consistency
  - PEG

Symptom Management

- Dysarthria and Communication
  - Therapy is rarely helpful
  - Speech therapist can help choose appropriate alternative communication methods
    - Writing with pen and paper
    - Alphabet boards
    - Electronic assistive communication devices that can be adapted for use with either hand or eye controls.

Symptom Management

- Dyspnea
  - Identify and treat reversible causes such as bronchospasm and pneumonia
  - Relaxation techniques
  - Psychosocial support
  - Modification in activity level
  - Use of a fan with cool air blowing on the face
  - Sit upright
  - Reassurance

Symptom Management

- Dyspnea
  - Noninvasive positive pressure ventilation
  - Systemic opioids are first-line agent
  - Benzodiazpines
  - Oxygen

Symptom Management

- Fatigue
  - Modify activities
  - There are medications that might be used to treat fatigue such as Modafinil or a two week trial of glucocorticoids

Symptom Management

- Muscle spasms
  - Mexiletine
  - Quinine (restricted by the FDA due to concerns regarding adverse effects)
  - Levetiracetam
  - Carbamazepine
  - Baclofen
  - Gabapentin
  - Tizanidine

Symptom Management

- Muscle weakness and functional decline
  - Assistive devices early in the disease
    - Canes, ankle foot orthoses, crutches, and walking frames
    - Wheelchair
    - Higher toilet seats
    - Bathtub lifts
    - Removable headrests
    - Specialized eating utensils, grips, and holders
    - Pressure-relieving mattress of air or high-density foam, along with proper positioning and repositioning to prevent pressure ulcers

Symptom Management

- Sialorrhea (Drooling)
  - Atropine
  - Hyoscyamine
  - Amitriptyline
  - Glycopyrolate
  - Botox
  - Low-dose radiation therapy to salivary glands if suggested if drooling does not improve with medication treatment

Symptom Management

- Thick Mucous
  - Increase fluid intake
  - Mucolytic (i.e acetylcysteine) if sufficient cough flow is present
  - Humidification of air
  - Cough augmentation with respiratory therapy

Symptom Management

- Pain
  - Caused by reduced mobility, muscle spasms or cramps, spasticity, and comorbid conditions.
  - Attentive nursing care is important
  - Frequent changes in position can help prevent pain, joint stiffness and skin breakdown.
  - Assistive devices such as special mattresses, pillows, and custom-fitted wheelchairs may help reduce pain
  - Treatment of muscle spasms and spasticity
  - Non-opioid analgesic medications
  - Opioids can be used when nonopioid analgesics fail.

Symptom Management

- Pseudobulbar affect
  - Dextromethorphan-quinidine
  - Tricyclic antidepressants: i.e. amitriptyline
  - SSRI's: i.e. Fluvoxamine

Symptom Management

- Monitor for symptoms of depression and get treatment when needed.

Symptom Management

- Sleep problems
  - Treat underlying causes
  - Nocturnal oximetry or sleep study can identify patients with disordered sleep patterns. They may benefit from noninvasive intermittent ventilation or NPPV
    - Bipap
    - CPAP
  - Sedatives
    - Use sparingly

Symptom Mangement

- Referral to Hospice
  - Recommended in the terminal phase of the disease
  - Discussion of advanced directives well in advance of the terminal phase and reviewed at least every six months
  - Work with Hospice team and Social worker.

Stephen Hawking

The End

Pete Frates

Ice Bucket Challenge

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