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Amyotrophic Lateral Sclerosis
August 23, 2019

Learning Objectives
After this course, participants will be able to:

- Describe at least three signs of ALS that resultant in loss of physical and cognitive functioning associated with the disease.
- Identify at least one environmental, social, and personal factor each that may impact function and participation in patients with ALS.
- Describe at least two theoretical approaches that will guide assessment and intervention for patients with ALS.
- List at least five common interventions and approaches to treatment used interchangeably by physical, occupational, and speech therapists.
Amyotrophic Lateral Sclerosis

- ALS, or Lou Gehrig’s disease, is a fatal, progressive neurodegenerative disorder resulting in profound muscle weakness and wasting of nearly all voluntary muscles, including the muscles needed for respiration.
- Amyotrophic=muscle atrophy – Lower motor neuron
- Lateral Sclerosis=degeneration and eventual death of motor neurons in the motor cortex, brain stem, anterior horn cells and pyramidal tracts – Upper motor neuron

Occurrence

- Occurs 1-2 per 100,000 persons worldwide (Redler & Dokholyan, 2012)
- More common in men until the age of 65
- Peaks between 40-70
- 5,600 diagnosed yearly in the US
- 5-10% of cases are familial (FALS)
- Median survival rate is 32 months from onset of signs and 19 months from diagnosis (del Aguila, et al., 2003)
Respiratory

- ALS causes weakness to the diaphragm muscle and other muscles needed for respiration.
- Respiratory failure is the most common cause of death (Pinto, et al., 1995).
- Mechanical Ventilation vs. palliative and hospice care

Pathophysiology of ALS

- TDP-43 – Transactive Response DNA Binding Protein 43)
  - Human cellular protein encoded by the TARDBP gene
  - Important role in normal neuron cellular functioning
  - Binds both DNA and RNA in neurons
  - Normally present within the cell body of neurons
- Excessive accumulation of TDP-43 has been found in the intracellular portion of motor neuron tracts in patients with ALS, and in the intracellular portion of neurons comprising the fronto-temporal lobe in patients with Fronto-Temporal Dementia (Mackenzie et al, 2010).
Pathophysiology of ALS

- **C9ORF72**: Gene located on chromosome 9
  - Associated with protein binding for axonal and dendrite growth
  - Found in the cytoplasm and presynaptic terminals of neurons
- Mutations to C9ORF72 have been linked to a diagnosis of ALS, FTD, and both ALS and FTD (Byrne et al, 2012; Ratti et al, 2012).
  - Lower age of onset for ALS
  - Impaired cognition
  - Impaired behavior
  - Abnormal changes in the grey matter of the frontal cortex
  - Reduced time of survival

Pathophysiology of ALS

- Molecular and genetic link between ALS and FTD have led some researchers to believe that the two diseases are a continuum of the same disease (Zago, 2011)
Pathophysiology of ALS

- **SOD1 Gene**: Superoxide Dismutase 1
  - Encodes the enzyme SOD1
  - Present on chromosome 21
  - The enzyme is an antioxidant that regulates superoxide, a byproduct of oxygen metabolism, that if left unregulated, causes cell damage
  - Mutations to the SOD1 gene have been linked with ALS
Pathophysiology of ALS

- Multiple theories on the cause of the sporadic form ALS:
  - Excess Glutamate – neurotransmitter
  - Neuro-inflammatory process
  - Environmental factors
  - Head trauma
  - Excessive physical activity
  - Genetic predisposition

Signs of ALS

- Onset presents with one or a combination of signs:
  - Lower Motor Neuron (LMN) involvement
  - Upper Motor Neuron (UMN) involvement
  - Bulbar Involvement
  - Cognitive Impairments
  - Respiratory Involvement
Upper and Lower Motor Neurons

- Lower motor neuron involvement causes:
  - asymmetrical muscle weakness and atrophy (Benny & Shetty, 2012)
    - Extensor muscles of the fingers and wrist
    - Intrinsic muscles of the hand (Thenar Muscles)
    - Dorsi-flexor weakness
  - Functional Impairments from LMN signs
    - Frequent tripping during ambulation
    - Difficulty buttoning shirts
    - Difficulty writing/typing
    - Difficulty holding and turning a key
Signs of ALS – LMN Continued

- Fasciculation
  - Small, localized, and involuntary muscle contractions
  - Normally occur in most humans due to spontaneous depolarization and repolarization, causing localized muscle twitching.
  - Fasciculations associated with ALS are pathological in nature due to damage to the LMN (Mills, 2010)

- Cramping
- Hypo-reflexia

Signs of ALS - UMN

- Spasticity
  - Occurs due to lesions in the UMN of the primary motor cortex (PMC)
  - PMC is responsible for voluntary and purposeful excitation and inhibition of LMN – resulting in coordinated muscle contractions and functional movement
  - Degeneration of neurons in the PMC causes loss of control of LMN
  - Causes difficulty with ambulation, ADL, and IADL (Ashworth, Satkunam, & Deforge, 2006).
Modified Ashworth Scale

<table>
<thead>
<tr>
<th>Grade</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>No increase in muscle tone</td>
</tr>
<tr>
<td>1</td>
<td>Slight increase in muscle tone, manifested by a catch or by minimal resistance at the end of the range of motion (ROM) when the affected part(s) is moved in flexion or extension</td>
</tr>
<tr>
<td>1+</td>
<td>Slight increase in muscle tone, manifested by a catch, followed by minimal resistance throughout the remainder (less than half) of the ROM</td>
</tr>
<tr>
<td>2</td>
<td>More marked increase in muscle tone through most of the ROM, but affected part(s) easily moved</td>
</tr>
<tr>
<td>3</td>
<td>Considerable increase in muscle tone, passive movement difficult</td>
</tr>
<tr>
<td>4</td>
<td>Affected part(s) rigid in flexion or extension</td>
</tr>
<tr>
<td>9</td>
<td>Unable to test</td>
</tr>
</tbody>
</table>

Signs of ALS - UMN

- Hyperreflexia
  - Increases sensitivity to deep tendon reflexes, including Babinski's sign (Chen et al, 2004)
    - Involuntary extension of first digit of the foot
Signs of ALS - Bulbar

- Bulbar signs are caused by deterioration of cranial nerves
- Facial weakness
- Tongue weakness
- Difficulty swallowing
- Dysphagia
- Dysarthria
- Sialorrhea (excessive production of saliva)

Signs of ALS - Cognition

- Fronto-Temporal Dementia FTD
  - Specific type of dementia
  - Differs from Alzheimer’s Disease (AD)
  - Often misdiagnosed as AD – despite clear differences
  - Behavioral Changes
  - Disregard for social norms
  - Apathy
  - Not associated with as much memory loss as AD
  - Patients with AD can often maintain social relationships, and do not have major behavioral changes, at least early in the disease
Signs of ALS - Cognition

- Cognitive Profile of ALS (in the absence of FTD) has been described as impairments in (Beeldman, et al., 2016): Meta analysis of the literature, controlling for FTD.
  - Speech Fluency
  - Language
  - Social Cognition
  - Executive Functions
  - Verbal memory

- Executive Functioning (Garcia-Madruga, Gomez-Veiga, & Vila, 2016).
  - Anticipation
  - Planning
  - Execution
  - Self-Monitoring

- Pseudobulbar symptoms
  - Inappropriate laughter
  - Inappropriate crying
  - Emotional Incontinence
Signs of ALS - Cognition

- Patients who test positive for executive function impairment at baseline have a faster rate of motor functional decline and death Elamin, et al. (2013).

Signs and symptoms

- Respiratory
  - dyspnea with exertion or lying flat
  - weak or ineffective cough
  - increased use of auxiliary musculature
  - daytime sleepiness
  - decreased concentration
  - headaches
ALS Usually Does Not Affect

- Bowel and Bladder
- Internal organs
- Sexual function
- Sensation - Posterior horn of spinal cord
- Eye musculature

What does a typical person with ALS look like?

This is a trick question—there is no standard presentation!
Diagnosis of ALS

- El Escorial Criteria – widely accepted as a method for diagnosing ALS (Brooks, Miller, Swash, & Munsat, 2000)

- El Escorial Criteria focuses on UMN and LMN signs and does not consider cognition.

<table>
<thead>
<tr>
<th>Clinical Presentation</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Definite ALS</strong></td>
</tr>
<tr>
<td>UMN and LMN signs in three regions (bulbar, thoracic, cervical, and lumbosacral).</td>
</tr>
<tr>
<td><strong>Probable ALS</strong></td>
</tr>
<tr>
<td>UMN and LMN signs in two regions AND some UMN signs must be rostral to (above) LMN signs</td>
</tr>
<tr>
<td><strong>Possible ALS</strong></td>
</tr>
<tr>
<td>UMN and LMN signs in one region OR UMN signs alone in two or more regions OR UMN signs alone in two regions</td>
</tr>
<tr>
<td><strong>Suspected ALS</strong></td>
</tr>
<tr>
<td>LMN signs alone in two or more regions</td>
</tr>
<tr>
<td><strong>ALL</strong></td>
</tr>
<tr>
<td>Signs of degeneration spread within or between regions AND lack of electrophysiological or neuroimaging evidence for other disorders which would explain observed degeneration</td>
</tr>
</tbody>
</table>
Onset of ALS

Riluzole (Rilutek)
- approved by the FDA for ALS treatment
- Increases survival by 2-3 months
- Symptom management

NPT520 (NeuroPore Therapies)
- Given orphan drug designation by FDA
- Penetrates blood-brain barrier
- Decreases neurotoxic misfolded proteins (superoxide dismutase 1) in the animal models
Treatment - Medication

- Tiglutik (Riluzole oral Suspension)
  - Same as rilutek
  - Liquid form vs. tablet to accommodate patients with dysphagia
  - Administered with an oral dosing syringe

- Radicava (Edaravone)
  - Removes free radicals
  - Provides neuro protection
  - Provides neuro protection in the absence of superoxide dismutase 1

Physical and Occupational Therapy

Goals for persons with ALS

- Maximize mobility and comfort through
  - Exercise modification
  - Stretching
  - Activity adaptation
  - Equipment prescription
  - Patient and family education
Exercise

- **Purpose:** Evaluate the effects of three different types of strictly monitored exercise programs versus a usual care program on retention of function in a cohort of ALS patients
- **Individual, Single Blind, Randomized Control Trial**
- **All participants had MMT of 3/5 or higher**
- **Intervention Groups:** 6 months
  - Active exercise performed with a PT, in a clinic plus a cyclogometer activity (20 minutes of upper and lower extremity cycle activity in a seated position).
  - Active Exercise without Cyclogometer
  - Passive Exercise protocol without active exercise
- **Measurement:**
  - Score on the ALS Functional Rating Scale (ALS-FRS). ALS-FRS is a validated rating instrument to monitor the progression of disability in patients with ALS. The ALS-FRS evaluated bulbar, motor, and respiratory function.
  - Forced Vital Capacity (FVC)
  - Perception of quality of life on the McGill Quality of Life Questionnaire (MGQoL)
  - All participants were measured prior to treatment, then monthly, until 6 months, then again at 6 months post treatment.


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Exercise

- **Primary Findings**
  - Exercise plus Cyclogometer group
    - Significantly higher ALS-FRS at the end of the 6 month treatment program and at the 6 month follow up, when compared to the “usual care” group.
    - All participants who underwent the active exercise program had higher ALS-FRS at the end of the 6 month treatment program and at the 6 month follow up, when compared to the patients in the “usual care group”
    - No significant differences in QOL were found
    - No significant rates of survival were found
Exercise

- Can be controversial in persons with ALS although some studies have shown benefits
- Is possibly detrimental in muscles that are exhibiting weakness
- If exercise is important to the patient, modify their program for safety and energy conservation

Exercise

- Stop if cramps or fasciculations occur
- Avoid eccentric contractions
- Avoid high resistance and high repetitions
- Monitor fatigue level
- Do not give resistive exercise to already weakened musculature
Stretch

- Hold 20-30 seconds
- Perform 3-5 repetitions daily
- Involve Caregivers
- We cannot increase muscle strength, but we can maintain muscle length with stretching
- Spasticity Management

Ecological Model of Human Performance

- Key Constructs of EHP
  - Person Factors include sensorimotor skills, cognitive skills, psychological abilities, personal interests, values, and experiences.
  - Task is an objective set of behaviors needed to engage in performance to reach a goal (Dunn, Brown, & Youngstrom, 2003).
  - Context includes temporal factors (age, lifecycle, health status, expectations), physical factors (objects and elements that surround us), social factors (relationships and norms), and cultural/environmental factors (beliefs and values).
  - Performance refers to the person’s engagement in tasks within a context.
Ecological Model of Human Performance

- Core Assumptions of EHP
  - It is impossible to examine a person outside of the context in which they live their lives (Dunn, Brown, & McGuigan, 1994).
  - Performance occurs as the person engages in a transaction with his or her context to engage in tasks. Combinations of specific tasks make up roles and occupations.
  - Contexts can enable and promote as well as obstruct and inhibit an individual’s performance and performance range (Dunn et al., 1994).
  - Performance Range is determined between the person and the context. The more effective the transaction between the person factors and contextual factors, the more extensive the performance range for the completion of tasks (Dunn et al., 2003).
  - Independence occurs when a person’s wants and needs are fulfilled. The EHP model views the use of assistive devices or support of others as resources or tools to achieve independence (Dunn et al, 2003).

(Dunn, Brown, & McGuigan, 1994).
Neuro-Functional Approach

- Theoretical framework originally designed for clients with traumatic brain injuries (TBI), who have severe cognitive, behavioral, and executive functioning deficits.

- NFA is a good fit with the ALS population, as they too can have severe motor, cognitive, behavioral and executive functioning deficits.

Neuro Functional Approach

- Theoretical Principles of the NFA (Giles, 2018)
  - Cognition and function are related, but the constructs of cognition or function are not reducible to the other.
  - One cannot assess functional performance abstracted from the context.
  - Skilled performance develops through attentive repetition of activities (neural restructuring).
  - The NFA was designed primarily for patients who are unlikely to spontaneously develop independence.
Neuro Functional Approach

- Assessment of patients using structured and unstructured observations in the natural environment.
- Interventions are targeted at the clients ability level, not the impairment level.
  - NFA recognizes that independence may not be restored in patients with ALS.
  - NFA approach with ALS patients involves ongoing activity and task modification and use of adaptive techniques and equipment.

Neuro Functional Approach

- Overlearning – deliberate over-practicing of a task past a set criterion (Driskell, Willis, & Copper, 1992)
- Errorless learning – Tasks are taught in a manner designed to eliminate the number of errors made during the learning phase (Masters, MacMahon, & Pall, 2004)
- Chaining techniques – Learning whole functional tasks by utilizing a step by step process to teach the sequential steps of a task until completion (Walls, Aane, & Ellis, 1981).
Summary of Theory

Mobility

- Cane
- Carbon Fiber AFO
- Rolling walker
- Companion wheel chair
- Power wheel chair
  - Tilt in space
  - Types of controls
  - Positioning recommendations
Mobility continued...

- Proper and safe techniques for transfers
- Family training
- Knowing when to limit or avoid ambulating
- Use of a Hoyer lift.

Activity Adaptation

- Energy conservation
- Home modification
- Driving
- Acceptance of help or use of devices
- Positioning to increase efficiency
Equipment Prescription

- ADL equipment—use only if time and/or energy efficient
  - Reacher
  - Button hook
  - Universal cuff
  - Sock donner—very limited population benefits from this

Button Hook
Tripod/Fine Motor Grasp

Cylindrical Grasp
Picture of feeding technique using proximal support

Computer Use

- Ergonomic Setup
- Voice typing software
- Onscreen keyboard with mouse use
- Eye gaze software
Equipment Prescription

- Assistive devices/transfer aides
  - Walkers—the ones with seats are not always useful
  - Canes—stick with single point canes
  - Transfer boards take too much energy in most cases
  - Pivot disc
  - Gait belt

Equipment Prescription

- Hand Splints
- Ankle orthotics
- Cervical collars
Equipment Prescription

- Wheelchairs
  - Companion
  - Manual wheelchairs—take too much energy
  - Don’t use insurance for scooters
  - Power wheelchairs
    - Tilt in space
    - Types of controls
    - Positioning recommendations

Pain

- Find the cause and treat it with
  - Stretching/ROM exercises
  - Positioning/supports (shoulder subluxation, digit or hand pain, back pain, cervical pain)
  - Modalities
  - Medication can help but has side effects
  - ALS is not a painless disease despite what you may read in the literature
Dysarthria

- Have the patient:
  - speak slowly
  - say the most important word first
  - write it down
  - use a communication board or device
  - answer yes or no question
  - spell things out

- Make sure they have a way to call for help if they cannot speak

Dysphagia

- Chew food slowly
- Avoid talking while chewing
- Avoid thin liquids
- Avoid foods with 2 different consistencies
- Try smaller, more frequent meals
- Peg Tube Considerations
SOB/DOE/Orthopnea

- Keep head of bed elevated.
- NIPPV(Bipap)—not oxygen!
- Avoidance of people with colds/respiratory infections

Secretions

- Medications
- Inexsuffulator
- Suction
Psychosocial Factors

- Depression: antidepressants and supportive therapy can help
- Insomnia: find out the cause and treat if possible
- Difficult decisions (vent/no vent, PEG/no PEG, where to live, who will be caregivers): Educating and empowering the patient to make a decision can help
- FTD can decrease decision making ability
- Financial Stress
- Access to Nutrition
- Utilities/Bills

Resources

- Government (Medicare, Medicaid, SSI, county)
- VA
- MDA
- Hospice
- ALSA
ALS Association

- Van transportation program
- Loaner “closet”: wheelchairs, tub benches, commodes, lift chairs, communication devices, etc.
- Accessibility program: help with ramps, stair glide rentals, emergency call devices
- Home Care: Supplement hours of care for patients
- Home Visits: Conducted by nurses, social workers, assistive technology specialist, mental health nurse, volunteers
- Resource Groups: for patients and caregivers

Questions?

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- University of Pennsylvania Health System
- Director of Rehab Services
- Penn Medicine at Home
- Scott.Rushanan@pennmedicine.upenn.edu
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